Giant Cell Arteritis

Roger E. Turbin, MD*
Mark J. Kupersmith, MD†

Address
*Department of Ophthalmology, Allegheny General Hospital, 320 East North Ave, Pittsburgh, PA 15212, USA
†Institute for Neurology and Neurosurgery, Beth Israel North, 170 East End Avenue at 87th Street, New York, NY 10128, USA

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Opinion statement

Diagnosis and management of giant cell (temporal) arteritis (GCA) should be performed by physicians who can accurately monitor the ophthalmologic, neurologic, and systemic sequelae of the disease as well as the numerous side effects of systemic corticosteroids, which are typically necessary for treatment. When the diagnosis of giant cell arteritis is seriously entertained, early treatment with adequate doses of oral or intravenous corticosteroids should not be delayed until laboratory confirmation has been obtained.

Unilateral or bilateral temporal artery biopsy should be performed on all patients with suspected GCA. A positive biopsy result mandates that higher doses of corticosteroids be used during the first 2 months, which comprise the critical period for risk of new ocular ischemia. A definitive, biopsy-proven diagnosis requires at least 6 months, and typically 12 months, of corticosteroid therapy.

Common pitfalls include increasing the dose and prolonging the use of corticosteroids in response to increases in the erythrocyte sedimentation rate (ESR) unrelated to GCA or visual blurring that may be related to benign tear film abnormalities, corticosteroid-induced lens changes, and other ophthalmic conditions. The muscle stiffness of polymyalgia rheumatica (PMR) must be distinguished from the osteoarthritis and other painful conditions common in the elderly.

After corticosteroid therapy has begun, continuing ophthalmologic evaluation is necessary to evaluate the effectiveness of treatment and whether ocular complications, such as glaucoma or cataract, develop.

Careful attention must be given to early detection and prevention of systemic side effects of corticosteroid treatment. Patients may be given gastrointestinal protective agents, such as histamine (H2)-blocking agents; vitamin D and calcium; oral hypoglycemic agents; and, if necessary, insulin and antihypertensive drugs. If bone density measurements warrant, hormones/supplementation to prevent or reverse osteoporosis may be prescribed.

After the initial diagnosis and first 4 weeks of treatment, elevation of the ESR or C-reactive protein alone should generally not be used as signs of disease activity nor as a reason to increase the daily dose of steroids. If symptoms or signs of disease activity occur, the dose should be raised regardless of test results. Even with vigorous physician-patient education, however, a patient is occasionally unable to provide adequate historical information about response to therapy, and the physician is forced to rely on laboratory values as a measure of disease activity.

After initial high-dose corticosteroid therapy, patients without a classic history and with negative biopsy results will generally receive a rapid taper to low doses of corticosteroids.

The role of repeated temporal artery biopsy in the clinical management of GCA is unclear. Despite persistence of PMR and, in some cases, histologic evidence of inflammation in temporal arteries, patients do not frequently have recurrence of symptomatic GCA after 6 months or more of corticosteroid therapy. Under these circumstances, late vision loss is rare.
Giant cell arteritis is a necrotizing granulomatous systemic arteritis with a predilection for arteries with elastic fibers in the intima, media, and adventitia. It is often threatening to sight and can be life threatening if the arteries of the kidney, heart, or aorta are affected \[1, \text{Class III}\]. Although a similar process may present in limited forms (e.g., PMR), such illnesses can evolve to the more serious GCA. Patients with PMR can progress to frank GCA even while being treated with low-dose corticosteroids \[2, \text{Class III}\]. Common signs and symptoms of GCA include headache (usually over the temporal fossa), scalp tenderness, temporal artery swelling and tenderness, jaw claudication, and anemia. Severe complications include blindness, ocular muscle ischemia, and cranial neuropathy. Less commonly, scalp and tongue necrosis, limb necrosis, myocardial infarction, arterial rupture, and aortic dissection occur. Steroid-responsive encephalopathy and transient ischemic attack of the vertebral-basilar distribution occur occasionally. Peripheral neuropathy, myelopathy, and multi-infarct dementia are rare. Despite some popular notions, carotid distribution infarcts caused by GCA are quite rare \[3, \text{Class III}\].

Signs and symptoms of vascular compromise in GCA are frequently subtle and are often present before visual loss is appreciated by the patient or an unsuspecting clinician. Signs of ocular and orbital ischemia can be identified and monitored with a comprehensive examination that includes dilated ophthalmoscopy, formal visual field examination, and ocular motility testing. Supplemental studies such as fluorescein angiography, ophthalmodynamometry, and color duplex ultrasonography of the temporal artery or orbit may also be helpful in the diagnosis.

Common ocular syndromes include central and branch retinal artery occlusions (Figs. 1 and 2), cotton-wool spots due to retinal ischemia (Fig. 3), anterior ischemic optic neuropathy (Fig. 4), choroidal abnormalities or infarction (Figs. 4 and 5), and posterior ischemic (retrobulbar) optic neuropathy, which can cause various visual field abnormalities and profound loss of visual acuity. Patients with posterior ischemic optic neuropathy may be the most difficult to diagnose because they present with visual dysfunction and an afferent pupillary defect in spite of an initially normal optic disc. GCA may be suggested by history, but other causes must be ruled out, including ophthalmic or carotid artery aneurysms, pituitary apoplexy, ischemic optic neuropathy, demyelinating disease, and non-GCA vasculitis.

In a patient with ischemic optic neuropathy or retinal infarction who is younger than 50, GCA is rarely found. In such cases, the clinician should consider other causes of vascular occlusion, such as Takayasu’s arteritis, Wegener’s granulomatosis, polyarteritis nodosa, systemic lupus erythematosus, and embolic disease.

**Table 1. 1990 criteria for the classification of giant cell (temporal) arteritis**

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<tr>
<th>Criterion</th>
<th>Definition</th>
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<tr>
<td>Age at disease onset ≥50 y</td>
<td>Development of symptoms or findings beginning at age 50 y or older</td>
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<td>New headache</td>
<td>Recent onset of or new type of localized pain in the head</td>
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<td>Temporal artery abnormality</td>
<td>Temporal artery tenderness to palpation or decreased pulsation, unrelated to arteriosclerosis of cervical arteries</td>
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<td>Elevated erythrocyte sedimentation rate ≥50 mm/h by the Westergren method</td>
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<td>Abnormal artery biopsy</td>
<td>Biopsy specimen with artery showing vasculitis characterized by a predominance of mononuclear cell infiltration or granulomatous inflammation, usually with multinucleated giant cells</td>
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*Traditional format.*